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Derzon to DHEW

ONCE AGAIN California and the West are sending an unusually able health professional to an important post in the government in Washington. Robert A. Derzon is a skilled and experienced administrator intimately familiar with the heartaches and triumphs of patient care, and the problems and challenges of education and research in the health sciences. He brings his intellect, his warm compassion and his considerable fiscal skills to the Health Care Financing Administration (HCFA), a newly created division of the Department of Health, Education, and Welfare (DHEW), as its first Administrator. He has stated:

> A real issue in this decade will be whether health professionals can set aside their self-interest to concentrate on the ways in which we can increase the health of our population. Hopefully, we can advise our sick patients to become prudent and judicious users of our expensive but vital hospitals and our health care resources. Government and the private sector must coalesce in finding ways to moderate the escalation in health care costs by assuring access to pluralistic and diverse health care systems, while maintaining high standards and humane service.

It is the interest of all concerned to achieve these goals. The nation should be well served with Bob Derzon in a position of leadership to accomplish them. He deserves and will need the help of us all.

—MSMW

Fibrosing Alveolitis

—*Extrinsic Allergic and Cryptogenic*

THE TERM "fibrosing alveolitis" has been coined by John G. Scadding of England as a generic name for the disease that had been described under such names as interstitial pneumonia or pneumonitis, acute or chronic Hamman-Rich disease or syndrome, idiopathic interstitial fibrosis, chronic diffuse sclerosing alveolitis, organizing interstitial pneumonia and usual interstitial pneumonia. Fibrosing alveolitis has been subdivided into two groups: extrinsic allergic alveolitis (typified by farmer's lung) and cryptogenic fibrosing alveolitis. In this country extrinsic alveolitis is referred to as "hypersensitivity pneumonitis"; whereas, cryptogenic fibrosing alveolitis is commonly called "idiopathic pulmonary fibrosis." A number of clinical, radiographic, immunologic and histologic features enable these two forms of "fibrosing alveolitis" to be clearly differentiated.[1]

Extrinsic allergic alveolitis is characterized by an acute or insidious onset of dyspnea, malaise, fever, muscle pains and weight loss. There is a history of frequent and regular inhalation of some organic allergen. Auscultation of the lungs may show fine crepitations. On roentgenograms of the chest, diffuse nodular infiltration with a tendency to involve mid- and upper lung fields may be seen. Fibrosis when it occurs mainly involves the upper lobes. Precipitating antibodies in the serum, belonging to immunoglobulins G (IgG) class, against the relevant allergen are usually present. Analysis of bronchial lavage fluid shows a striking increase in T-lymphocytes and immunoglobulin M (IgM) along with some eosinophils and IgG.[2] In early disease histologic examination of the lung tissue shows noncaseating granulomata, but in the advanced stage, granulomata are replaced by fibrous tissue. Corticosteroids are effective in the early stage before fibrosis is established.

Cryptogenic fibrosing alveolitis or idiopathic pulmonary fibrosis differs from the extrinsic type in many ways. Dyspnea on exertion is the constant symptom but fever, muscle pains and weight loss are absent. Clubbing of the fingers and toes occurs in as many as 70 percent of the patients and crepitant rales are present in more than two thirds of the patients. Radiological appearance varies with the stage and extent of the disease and includes ill-defined patchy opacities, nodular infiltrates, linear and reticular shadows, "ground glass" appearance and honeycombing. Unlike extrinsic allergic alveolitis the roentenographic abnormality is diffuse. Lung function studies show reduction of static lung volumes and diffusing capacity for carbon monoxide but the degree of fibrosis is best correlated with the change in arterial oxygen tension and alveolar-arterial oxygen difference on exercise and the coefficient of lung retraction (maximum static transpulmonary pressure/observed total lung capacity).[3] Immunologic

studies show clear evidence of a T-cell mediated process against collagen in this disease.[4] In about two thirds of the patients, antinuclear factor, or rheumatoid factor, or both, are present in the serum. Furthermore, cryptogenic fibrosing alveolitis is accompanied in a proportion of patients by certain other autoimmune disorders; for example, Sjögren syndrome, Hashimoto thyroiditis, chronic active hepatitis, polymyositis and hyperglobulinemic renal tubular acidosis.[5] The occurrence of familial cases and some preliminary data indicating an increase in HLA-12 and HLA-29 in patients suggest that there may be a genetic predisposition to cryptogenic fibrosing alveolitis.

The histopathologic features can be described in a range between two extreme patterns which may be called cellular and fibrotic (Scadding and Hinson prefer terms desquamative and mural, respectively[6]). The cellular pattern is characterized by the presence of inflammatory cells mainly lymphocytes, macrophages and plasma cells. Eosinophils and neutrophils are also noted. The fibrotic pattern is distinguished by the prominence of alveolar wall thickening and fibrous tissue with fewer inflammatory cells. Recent studies suggest that the process starts as an alveolitis and progresses to interstitial fibrosis.[4] Once the disease has reached the fibrotic or mural stage it probably becomes irreversible, but the cellular or desquamative stage can be successfully treated in many cases with corticosteroids. Immunosuppressive drugs have been found useful in some cases but their role in the management of cryptogenic fibrosing alveolitis remains uncertain.[7]

In all cases of fibrosing alveolitis or diffuse interstitial fibrosis the possibility of extrinsic allergic alveolitis should be considered. Once the diagnosis is firmly established, institution of corticosteroid therapy and avoidance of offending allergen can bring the favorable outcome. If the clinical, radiographic and immunologic features point to the diagnosis of cryptogenic fibrosing alveolitis, a lung biopsy may be justified to stage the disease because corticosteroids if given in cellular or desquamative cases may prevent formation of fibrosis. Further clinical trials, however,[7] are needed to establish the value of lung biopsy as a guide for selecting therapy in patients with cryptogenic fibrosing alveolitis.

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